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AUTOIMMUNE POLYGLANDULAR SYNDROME (TYPE III C) AND NON-SUSTAINED VENTRICULAR TACHYCARDIA – A RARE CASE REPORT

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Abstract

Failure of two or more endocrine glands together requiring hormone replacement is termed as "Autoimmune polyglandular syndrome" (APS). The aim of this case report is to underline the possible etiological link between primary hypothyroidism and non-sustained ventricular tachycardia (NSVT) followed by ventricular premature complexes (VPC) and ventricular trigeminy, although supraventricular arrhythmias are common features of hyperthyroidism. Here we present a case of 47 years old patient with repetitive episodes of ventricular premature complexes, with autoimmune thyroiditis that was diagnosed 3 years ago, and was on thyroxin replacement therapy, which could be an attributed risk for reactivation of arrhythmia.

Introduction

Autoimmune polyglandular syndrome (APS) is associated with two or more endocrine disorders, which are mediated by autoimmune mechanisms and lead to a hypofunctional state. Due to autoimmune pathogenic mechanisms, APS has become an increasingly recognized clinical entity in endocrinology. Patients with APS are currently treated with appropriate replacement therapy for each deficient endocrine organ system.

In the future, early recognition of these disorders and delineation of their cause and pathophysiology may help to improve the clinical scenario and prevent other systemic deterioration. Recent literatures suggest that the initiation of Thyroid supplementation in case of hypothyroidism should be done after ruling out any underlying cardiac conditions, as thyroid hormone replacement therapy can precipitate arrhythmias in an undiagnosed cardiac case.

In this paper, we report an emblematic case of APS type 3c (details classifications are discussed below) with NSVT followed by ventricular premature complex and ventricular trigeminy, features not described in previous literatures.

Case Report

A 47 years old gentleman, presented to the emergency room with complaints of acute onset palpitations and chest pain for 1 day, with preceding history of dyspnea and dizziness for 4 days. He was a non-smoker, non-hypertensive, and no history of bronchial asthma, or any thromboembolic events (DVT, pulmonary embolism, stroke, MI, amaurosis fugax, limb ischemia) in the past. One year ago he was known to have premature ventricular complexes and a 24 hour Holter monitoring revealed frequent ventricular ectopics in form of isolated beats, couplets and triplets with occasional supraventricular ectopics, with no episode of supraventricular tachycardia, non sustained or sustained ventricular tachycardia, for which he was conservatively treated on Tab. Amiodarone 200 mg twice daily for 2 weeks. Patient is a known case of type II diabetes mellitus for the past 8 years and was on oral hypoglycemic agents for the same. Simultaneously, patient is a diagnosed case of primary hypothyroidism for 4 years for which he was on replacement therapy (50mcg once daily). Patient also presented with concerns of alopecia areata, dry scaly skin and vitiligo in the sun-exposed area for which he sought treatment, which however did not subside. Detailed history taking revealed positive family history of chronic thyroiditis with hypothyroidism in the mother and sister, and father had diabetes mellitus type 2.

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General examination of the patient showed a conscious, oriented and afebrile patient with heart rate 72/min, B.P-128/98 mmHg, respiratory rate 16/min and SpO2 – 94% on room air. Chest examination and other systemic examinations were unremarkable. Local examination unveiled hypo-pigmented patches in the hands, feet and arms with patchy hair loss and dry scaly skin over the arm and forearm.

Initial blood investigations reported normal haemogram, kidney/liver function tests, cardiac biomarkers and arterial blood gas analysis. Chest X-ray was within normal limits. Old ECG showed monomorphic non-sustained ventricular tachycardia and serial ECG at our center revealed frequent premature complexes (VPC), couplets and occasional run of ventricular trigeminy (Fig 1 -2). 2D echo revealed no regional wall motion abnormality with LVEF – 60%.

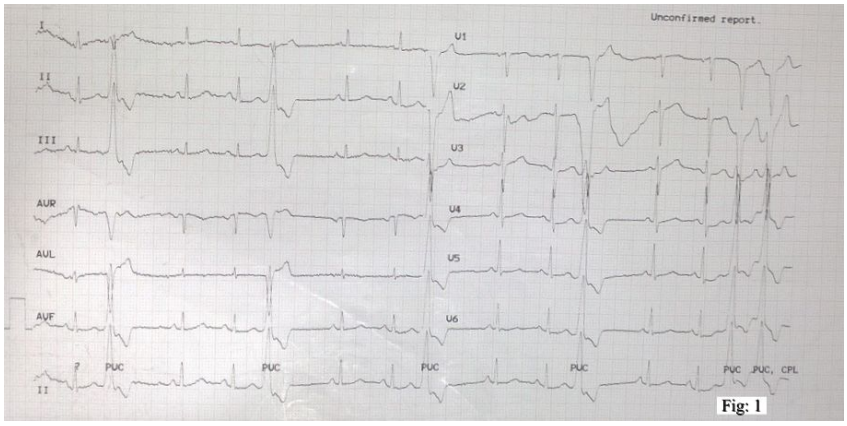

Fig: 1 – Ventricular trigeminy and couplets

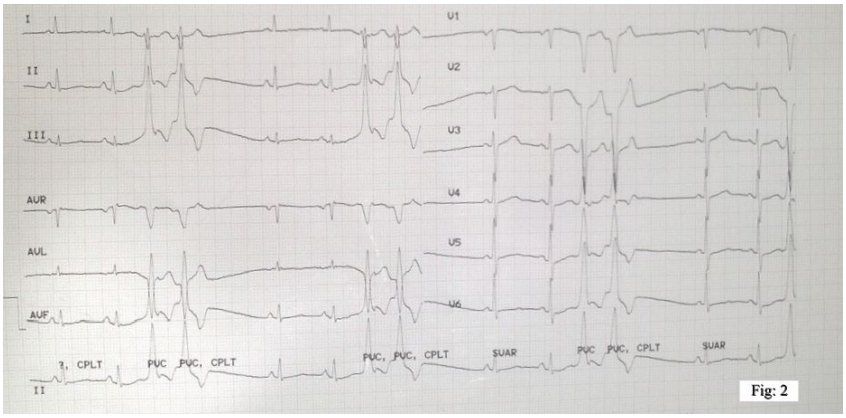

Fig: 2 –Frequent VPCS and couplets

The patient was admitted in ICCU for observation and close vitals monitoring. His initial treatment modality included Amiodarone infusion, dual anti platelets, statins and Inj. NTG 0.3ml/hour infusion.

Thyroid function was investigated which revealed T3 – 2.09 pg/mL (2.00-4.40), T4-0.81pg/mL (0.93-1.70), TSH-39.59 uIU/mL (0.27-4.20) and anti thyroid peroxidase antibody more than1000 IU/ml (0.00-5.61).

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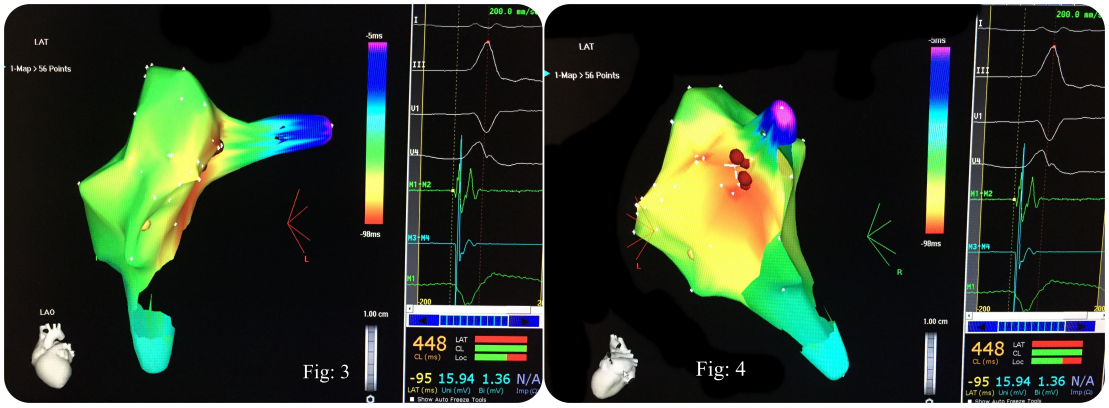
Coronary angiogram was performed to rule out ischemia as the cause of ventricular ectopics and findings were suggestive of non-critical coronary artery disease. Electrophysiological studies showed frequent outflow tract PVCs arising from right ventricle. The VPCs were mapped on 3D mapping (Carto) and were localized to postero septal area of right ventricle outflow tract. Successful radiofrequency ablation at the site resulted in absence of any further VPCs. 2D echo done post procedure had no significant changes.



(*Fig: 3-4 VPCs were mapped on 3D mapping (Carto) (LAT view and LAO view) and were localized to postero septal area of right ventricle outflow tract*)

Patient's clinical condition improved progressively, and was discharged on single antiplatelet drug, statin and thyroid replacement therapy.

Discussion

Multiple endocrine gland insufficiency that is associated to an autoimmune disease was initially termed as Autoimmune Poly endocrine Syndrome

(APS). However in 1980, Neufeld and Blizzard suggested a classification of APS, based on clinical criteria only, and further described them into four main types. (1,2)

- APS-1: chronic candidiasis, chronic hypoparathyroidism, Addison's disease (at least two present).
- APS-2: Addison's disease (always present) + autoimmune thyroid diseases and/or type 1 diabetes mellitus.
- APS-3: Autoimmune thyroid diseases associated with other autoimmune diseases (excluding Addison's disease and/or hypoparathyroidism)
- APS-4: Combinations not included in the previous groups

APS-3 was further sub classified into various entities based on the association of diabetes mellitus, pernicious anemia, vitiligo, alopecia, hepatic autoimmune disease excluding adrenal involvement, which is simplified and depicted in the table (Fig: 1)

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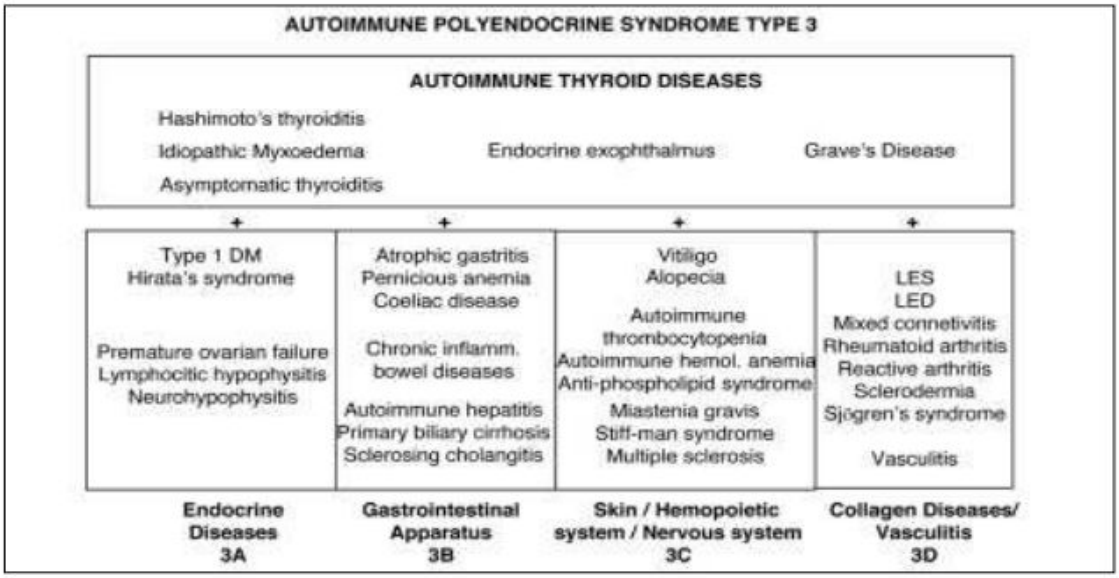

Gherbon Adriana ET all demonstrated the prevalence of autoimmune chronic thyroiditis (ACT) in DM type 2 was 26.55% (77 patients, 69 F and 8 M). In adults with DM type 2 APS type III was found in 6 (2.06%) cases, of which all have ACT and vitiligo. In these cases the patients with autoimmune disease could have been latent autoimmune diabetes in adults (LADA). Therefore, if we have a patient with two or more autoimmune disease, we should investigate this for another possible autoimmune disease. (3)

Classical ECG manifestations in a patient with primary hypothyroidism are (4):

- Prolonged conduction defects
- Low voltage ECG
- Sinus bradycardia
- Atrio-ventricular or bundle branch blocks.

NSVT can also be part of the cardio-vascular anomalies during primary hypothyroidism as seen in our case, though few literatures have reported this.

Tachyarrhythmia in primary hypothyroidism could be due to the following causes (4):

- Alteration of myocyte-specific gene expression
- Interstitial edema
- Myofibril swelling with loss of striation
- Increased arterial stiffness
- Endothelial dysfunction
- Premature atherosclerosis
- Disturbances of the sympathetic-vagal tone with a relative increase in sympathetic tone and autoimmunity.

So far, no case of APS type 3C presenting with non-sustained ventricular tachycardia was found in our search of literature.

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This patient presented with non-sustained ventricular tachycardia followed by frequent premature complexes (VPCs), couplets, occasional run of ventricular trigeminy and co-existing autoimmune thyroiditis along with diabetes mellitus, alopecia areata and vitiligo (APS IIIc), in which systemic problems were managed accordingly, with arrhythmia control as our top most priority. During replacement therapy, continuous monitoring of thyroid function is required to avoid over replacement as it can lead to premature osteoporosis and cardiac arrhythmias. (5, 6)

Conclusion

The presence of an autoimmune endocrinopathy urges the need to investigate other endocrine dysfunction in order to decrease the cardiac morbidity and mortality. Early recognition of such cases and replacement therapy could be lifesaving, bearing in mind the regular monitoring of thyroid function test to avoid iatrogenic adverse effects. In cases with life-threatening arrhythmia we advocate appropriate treatment of the arrhythmia in top priority.

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Conflicts of interest

No potential conflict of interest relevant to this article was reported.

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